CASE REPORT

Keratinous inclusion cyst of oesophagus: unusual finding

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SUMMARY

Cysts of the oesophagus are unusual findings and they are classified according to the embryological site of origin. It may represent inclusion cysts, retention cysts and developmental cysts. We present a case of keratinous inclusion cyst of the lower oesophagus in a 71-year-old Malay woman who presented with dyspepsia and severe epigastric pain. An oesophago-gastro-duodenoscopy demonstrated a sliding hiatus hernia with whitish ulcer-like lesion at the lower oesophagus. Biopsy from the lesion revealed a keratinous inclusion cyst. The patient was given pantoprazole and put on regular follow-up for monitoring any other development.

BACKGROUND

Cysts within the oesophageal wall may represent inclusion cysts, retention cysts and developmental cysts.¹ Inclusion cysts may be lined by squamous, columnar or ciliated epithelium. Retention cysts or mucocoeles arise from cystic dilation of submucosal glands. Developmental cysts are also known as duplication cysts of oesophageal, bronchial or gastric origin. Keratinous inclusion cyst in the oesophagus is a new observation because the cyst is lined by squamous epithelium with distinct granular layer and the lumen contains lamellated keratin. Keratinous inclusion cysts normally occur in the skin and result from traumatic inclusion of the epidermis. The aim of this report is to present a rare case of keratinous inclusion cyst of the oesophagus and review of the literature.

CASE PRESENTATION

A 71-year-old Malay woman with background of diabetes mellitus and hypertension for 20 years was admitted to the medical ward with severe epigastric pain for 3 days. She described the pain as burning in nature and radiated to the back. It was associated with nausea, recurrent vomiting and poor oral intake. The pain was not related to food. She denied any history of fever, haematemesis, passing blackish stool, shortness of breath and symptoms of obstructive jaundice. On further questioning, the epigastric pain started 5 years ago with frequent visit to private clinic and hospital and was treated with antacid. She was a non-smoker and non-alcoholic.

On physical examination, she was alert and conscious with respiratory rate of 20/min and oxygen saturation of 100% under room air. Her blood pressure was 139/90 mm Hg and heart rate was

90 bpm. Abdominal examination revealed mild epigastric tenderness and no hepatosplenomegaly.

INVESTIGATIONS

A series of blood investigations such as full blood count, coagulation studies, renal profile, serum amylase and cardiac enzymes were normal. The patient was subjected to oesophago-gastro-duodenoscopy (OGDS) in view of recurrent epigastric pain. OGDS finding showed sliding hiatus hernia with lower oesophageal ulcer-like lesion with evidence of severe reflux oesophagitis (figure 1). Biopsy of the lower oesophagus was taken and sent for histopathological examination.

Grossly, there were only two tiny fragments of grey-whitish tissue measuring 4 mm each. On microscopic examination, serial sections showed fragmented stratified squamous epithelium without the underlying stroma (figure 2). Within the subepithelial mucosa, there was a cystic lesion (figure 2) lined by stratified squamous epithelium exhibiting granular cell layers (figure 3). The lumen of the cyst was filled with keratin flakes (figure 3). No muscular layer or cartilaginous tissue was seen.

TREATMENT

The patient was prescribed with pantoprazole 40 mg twice daily and was advised to attend the surgery outpatient clinic.

OUTCOME AND FOLLOW-UP

The patient was responding to the prescribed medication and on regular follow-up.

DISCUSSION

Oesophageal cysts are rare. It accounts for about 10–15% of cases in all primary masses of the mediastinum.² They are classified according to embryological site of origin. It may represent inclusion cysts (lined by squamous, columnar or ciliated epithelium), retention cysts or mucocoeles and developmental or duplication cysts.¹ A few authors divided the oesophageal cysts into congenital and acquired cysts. The congenital oesophageal cysts include duplication cysts, bronchogenic cysts and inclusion cysts in the congenital group.^{3–4} Acquired cysts include oesophagitis cystica (cystic growths developing from the submucosal gland within the upper third of oesophagus) and paraoesophageal cysts (a round regular mass in the posterior mediastinum).⁵

Inclusion cysts of oesophagus are intramurally located and have epithelium of either respiratory or squamous type. They cannot be called duplication or bronchogenic cysts as they are not covered by

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Figure 1 Clockwise from top left: gastro-oesophageal sphincter area shows lower oesophageal ulcers with severe reflux oesophagitis (arrow). Normal fundus, duodenum and antrum.

two muscle layers and do not contain cartilage. Owing to uncertainty of aetiology, the term inclusion cyst is used. Keratinous inclusion cyst is a new observation wherein the cyst is lined by squamous epithelium with lamellated keratin within the lumen. There was one case report found in the literature, by Thomas *et al*,⁶ describing an upper oesophageal sebaceous cyst in a 61-year-old white woman who presented with chronic sore throat for 1 year duration. The frequent use of name sebaceous cyst for keratinous cyst is a misnomer as it is based on the gross appearance of the cyst content. Sebaceous cyst in the oesophagus is thought to arise from heterotopic sebaceous glands in the oesophagus by other workers but Thomas *et al*⁶ considered them to originate from pluripotent cells in the oesophageal mucus glands. The cyst we report here is not related to the

sebaceous gland or the sebaceous cyst. Therefore, the sebaceous cyst of the oesophagus reported by Thomas *et al* was different from our case.

Vougiouklakis *et al*⁵ reviewed a large series of autopsies for oesophageal cysts in order to establish the incidence. They found six oesophageal cysts out of 49 196 autopsy cases, giving the incidence of 1 in 8200.²⁻⁴ They also reported an inclusion cyst of oesophagus in a 41-year-old man who presented with dysphagia and regurgitation. OGDS showed a polyp-like mass in the proximal third of the oesophagus and biopsy revealed an inclusion cyst.⁵ However, the cyst reported here was lined by cubocolumnar ciliated epithelium, not a squamous epithelium as in our case.

Keratinous inclusion cysts represent the most common cyst of the skin. Historically, these cysts have been referred to by

Figure 2 The lower oesophageal mucosa displays fragmented non-keratinising parakeratotic stratified squamous epithelium. Beneath the mucosal epithelium, a keratinous cyst is seen (H&E stain, $\times 4$).

Figure 3 The cyst is lined by stratified squamous epithelium with the presence of prominent granular layer. The lumen of the cyst is filled with flakes of lamellated keratin (H&E stain, $\times 10$).

various terms, including follicular infundibular cysts, epidermal cysts, epidermoid cysts, keratin cysts, sebaceous cyst and epidermal inclusion cysts. The term epidermal inclusion cyst refers specifically to an epidermoid cyst that is the result of the implantation of epidermal elements in the dermis. Although named an epidermal inclusion cyst, the inclusion itself sits within the dermis and results from the proliferation and accumulation of keratin into a cystic mass that invaginates into the dermis. The inclusion cyst may also arise secondary to a traumatic event that leads to implantation of the epidermis into the dermis. The term sebaceous cyst is better be avoided because it is a misnomer implying cyst is of sebaceous origin, which it is not.^{7 8} The occurrence of this type of cyst within the oesophagus was very unusual and the pathogenesis need to be explored.

In general, the usual clinical course of an oesophageal cyst is benign and many patients remain symptom free. Surgical resection is the treatment of choice even when the lesion is asymptomatic. Enucleation and poypectomy by oesophagoscope represents a useful non-surgical alternative treatment for cysts accessible through the endoscope.³ An untreated cyst may enlarge and may be complicated with intracystic haemorrhage, perforation or infection.⁹ Malignant change of cyst has also been reported. McGregor *et al*² reported the first case of squamous cell carcinoma arising from an oesophageal intramural inclusion squamous epithelial cyst in a 65-year-old black man.

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Competing interests None.

Patient consent Obtained.

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Learning points

- Oesophageal cysts are divided into three groups, which are inclusion cysts, retention cysts or mucocoeles and developmental or duplication cysts.
- Keratinous inclusion cyst commonly occurs within the dermis and the findings within the oesophagus are a new observation.
- Keratinous inclusion cysts are lined by squamous epithelium with presence of granular layer and the lumen filled with lamellated keratin.

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